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Angiomatoid Fibrous Histiocytoma of the Arm Treated by Radiotherapy for Local Recurrence—Case Report

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The Angiomatoid Fibrous Histiocytoma is a rare tumor of soft tissues, which occurs mainly in children and young adults, with low malignancy grade. It has the capacity of local recurrence, but rarely metastizes. It is frequently difficult to differentiate this from vascular tumors, namely hemangioendotheliomas and angiosarcomas, or simply organized hematomas. The authors present a case of a patient with an angiomatoid fibrous histiocytoma of the arm, treated with radiotherapy after three postsurgical recurrences. Med. Pediatr. Oncol. 28:373–376, 1997. © 1997 Wiley-Liss, Inc.

Key words: angiomatoid fibrous histiocytoma; radiotherapy

INTRODUCTION

The broad group of the so called ''fibro-histiocytic'' tumors of soft tissues, involves a large variety of mesenchymal neoplasms with great clinical, morphological, and behaviour heterogeneities [1,2,3]. So, there is much concern in trying to identify in this group, variants that may constitute individualized anatomo-clinical entities, since they have similar manifestations, treatment, and prognosis [4,5]. One of these entities, individualized in the new classification proposed by the WHO for Soft Tissue Tumors, is the Angiomatoid Fibrous Histiocytoma (AFH), described by Enzinger in 1979 under the designation of Malignant Fibrous Histiocytoma, angiomatoid variant [3].

It is a rare neoplasm, which occurs mainly in children and young adults, with low malignancy grade, with the capacity of local recurrence but rarely metastizes, being found usually in the limbs and with histological characteristic features, although it frequently raises differential diagnoses with hemangioendothelioma-like vascular tumors or simply organized hematomas [2,3].

The AFH usually manifests itself by a slow growing superficial nodule which the patient notices only after some time of evolution. Macroscopically, the tumor is circumscribed, multinodular, sometimes with imprecise limits, whose cut surface is brownish, cystic, and hemorrhagic due to its great vascularization [3,6,7].

The histology has its own features, well defined, fibroblast-like spindle cells with round or oval nucleus, abundant cytoplasm, myofibroblast-like, and sometimes with hemosiderin [1,2]. The similarity of these cells with histiocytes, is the origin of the name of histiocytoma that the whole group of these neoplasms are called, even though today we know that they have nothing to do with the monocityc-macrophagic system and, much the opposite, they show immunocytochemical and ultrastructural markers like young myofibroblasts, with little differentiation [3]. The tumoral cells are organized in solid nodules or in bundles that limit cystic spaces and may look like vessels, moreover since these tumors are very vascularized and hemorrhagic. Peripherically, where the growth pattern is usually infiltrative, it is common to find fibrosis and other areas of high inflammatory infiltration of lymphoplasmocytic cells, sometimes even organized in lymphoid follicles. Occasionally there are so many lymphocytes that the lesion may simulate a lymph node metastasis [2,3,5].

We present a case of a patient with an AFH of the arm, who was treated successfully with radiation therapy after three postsurgical recurrences.

CASE REPORT

The patient was a 6-year-old caucasian boy (at the time of diagnosis), who was presented in February 1985, with a complaint of a 0,5cm nodule, in the upper third of the medial face of the right arm, regular, without pain or inflammatory signs. There was no history of prior trauma. In November 1985, there was an increase in the size of the nodule. He was observed in his local hospital

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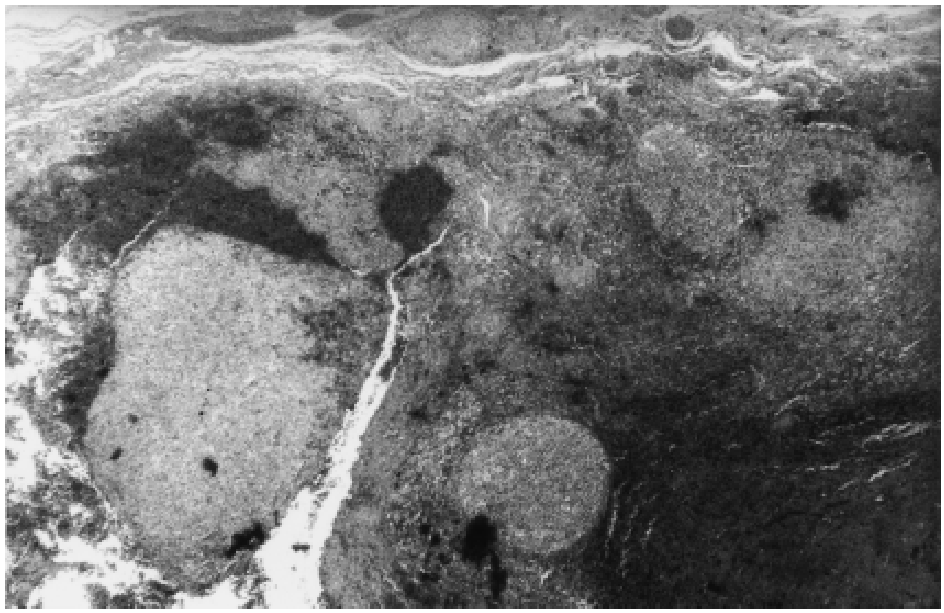

Fig. 1: Angiomatoid fibrous histiocytoma, showing tumor mass surrounded by a dense fibrous pseudocapsule with hemorrhagic spaces and prominent lymphoid cuff (×40).

and underwent incisional biopsy which was diagnosed as *''rhabdomyosarcoma''*. Surgical excision of the nodule was proposed, but it was not performed because of cardio-respiratory arrest during surgery.

Referred to our institution, he was first observed in March 1986. At this time, he was asymptomatic, with a good performance status. Physical examination on admission showed only the nodule already described, now measuring 1,0cm in diameter. Results of admission laboratory studies, including liver function tests, were within normal limits. Chest radiograph and bone scan were normal.

He underwent total excision of the nodule in March 1986. The surgical exploration revealed a pulsatile, reddish mass, highly vascularized, probably related to the vessels. The nodule was 3,5cm in diameter, cystic, encapsulated, and hemorrhagic. The diagnosis of angiomatoid fibrous histiocytoma was made on the basis of the histological features (Fig. 1), and ultrastructural studies.

Three years postoperatively, the patient developed a nodule, 1,0cm diameter, in the same site as the previously excised lesion, which was painful and thought to be a probable recurrence.

He underwent total excision of the second nodule in July 1989. The histologic features confirmed recurrence of an angiomatoid fibrous histiocytoma.

Six months after the second surgery a nodule, 2,5cm diameter, was found again, in the same place. This time, he underwent a partial excision, because the tumor extended to the humeral artery. The histologic features revealed an angiomatoid fibrous histiocytoma, with an increase of cellularity and mitosis (Fig. 2).

The arteriography of the right arm revealed normal vascularization, with a small blush in the terminal branches of the cubital artery.

A course of radiation therapy was designed, with the intention of curing the disease as well as preserving limb function. Therapy was divided into two parts, with no period of rest in between.

In the first portion of radiation therapy, the right arm-excluding the proximal and distal epiphysis of the humerus, in order not to impair bone growth-was treated in two fields using anterior and posterior opposed portals. These fields received 50,4Gy in 28 fractions, five times a week, via a 6MV photon linear accelerator beam. In the second portion, a boost to the surgical area was performed. He was treated with a direct anterior field, which received 16Gy in eight fractions, via a 6MV photon linear accelerator beam.

The patient tolerated the treatment reasonably well, with no loss of function in the affected extremity. Further studies revealed complete resolution of the lesions, and there has been no clinical or radiological evidence of tumor recurrence seven years after radiotherapy.

DISCUSSION

The AFH is a rare subtype of fibrous histiocytoma with low-grade malignancy, with its own anatomo-clinical characteristics such as clinical findings, histological features, and an excellent prognosis, that justify its individualization [2,3,4,8,9].

Their cells frequently show myofibroblastic and his-

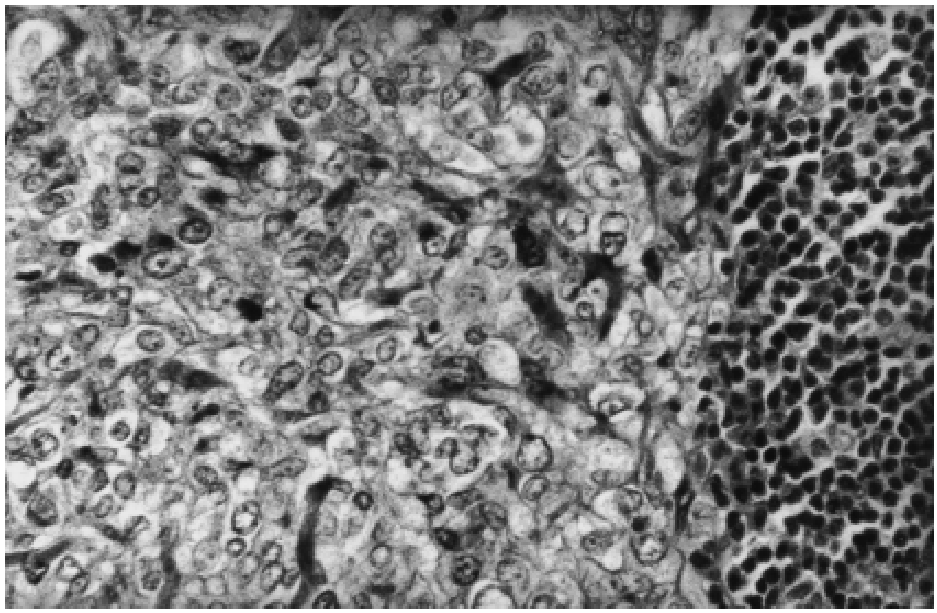

Fig. 2: Cellular area from an angiomatoid fibrous histiocytoma. Histiocyte-like cells are arranged in solid sheets. Lymphoid infiltrate surrounds tumor nodule (×400).

tiocityc appearance and occasionally, they present high phagocytic activity [6,7]. Their rich vascularization and its architectural pattern, may cause differential diagnostic difficulties with other vascular tumors, namely hemangioendothelioma and angiosarcomas. In a study from Smith *et al*, which included 19 cases of AFH, immunohistochemistry was used for the assessment of the histiocytic antigens CD68, L1, factor XIIIa, and S100 protein and for keratin, leukocyte common antigen, factor VIII-related antigen, muramidase, and desmin on six of these cases. Nine of 19 cases expressed the histiocyte differentiation antigen CD68, and in three cases, expression was seen in more than 90% of cells. Of the 19 cases studied, three contained rare S100-positive tumor cells. Two of them expressed factor XIIIa within tumor cells. In three of the six cases, rare desmin-positive tumor cells were identified, a fact that indicates their myofibroblastic nature. Neoplastic cells did not express the other antigens studied, namely factor VIII, which is frequently found in vascular tumors. Immunohistochemical findings were not in agreement with conventional malignant fibrous histiocytoma features, and this supports the fact that its angiomatoid form is a distinct entity [4].

Enzinger states that three features, occurring in varying proportions, contribute to define this lesion: a) irregular solid masses, made of fibroblasts, myofibroblasts and histiocyte-like cells b) focal areas of hemorrhage or cyst-like spaces c) chronic inflammatory cells, namely lymphocytes and plasma cells [2,3].

In general, the cells interspersed with areas of hemorrhage occupy the central portion of the tumor. The inflammatory cells may also be present in the center of the nodule, but mainly they form a dense peripheral cuff that blends with the surrounding pseudocapsule [1,2,3].

In the same study, follow-up information was available in 24 of the 41 patients. Eleven had one or more recurrence, with a disease-free-survival less than 12 months. Three patients died in consequence of metastatic disease, that was disseminated in two cases and pulmonary in the other. In all patients, the treatment consisted only in surgical excision of the tumor [2].

Despite the resemblance of some of these lesions to a vascular tumor, such as hemangioendothelioma, the hypothesis of its vascular origin seems to be remote. In fact, in the original report from Enzinger, most of the large cyst-like hemorrhagic spaces were circumscribed by several layers of fibrohistiocytic cells, often with considerable deposits of hemosiderin in their cytoplasm, suggesting focal hemorrhage rather than a vascular neoplasm. Moreover, the negativity of the neoplastic cells to endothelial markers, also supports the fibro-histiocytic origin, and not vascular, of these tumors [2,3,5].

On the basis of the original report from Enzinger, Costa *et al* reported a study of 108 new cases of AFH, based on its clinical and pathologic features and follow-up, to determine the long-term behaviour of the neoplasm, and whether various histologic features, such as atypia, mitosis, infiltrative borders after surgery, and inflammatory infiltrate are useful in predicting outcome. The age ranged from 2 months to 70 years, with a median age of 14 years. Females (55%) predominated over males (45%). The tumor occurred most commonly on the extremities (65%) followed by the trunk (28%) and the head and neck (7%). Eleven patients developed local

recurrences, related to incomplete excisions, with a mean period of 7 months after treatment. Likewise, the anatomic location of tumor had influence, with a greater local recurrence rate in the region of the head and neck. This is likely due to the difficulty of performing a wide local excision in this area. Five patients developed metastatic disease, four of them presenting local metastases, whereas the fifth patient developed pulmonary and cerebral metastases. The other parameters evaluated (mitotic activity, pleomorphism, inflammatory infiltrate, size of tumor, and age), were not related to its clinical behaviour [1].

The treatment of choice is still surgical, whenever possible. In face of these results, it seems important to continue to evaluate the role of radiotherapy and chemotherapy.

In our case, the role of radiotherapy was fundamental in the local control of the tumor.

However, further studies are necessary in order to make definitive conclusions.

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